

**Supplementary Table 1:** Clinical and immunological characterization**Clinical manifestations**

Respiratory tract infections	Recurrent pansinusitis Recurrent pneumonia Bilateral bronchiectatic disease
Other infections	Cholecystitis
Autoimmune disease	No clinical evidence
Granulomatous disease	No clinical evidence
Lymphoproliferation	No clinical evidence
Enteropathy	No clinical evidence
Age of onset	Child

<b>Immunological features</b>	<b>Value</b>	<b>Reference values</b>	<b>Year</b>
<i>Immunoglobulins</i>			
IgG	3 g/L	7-16 g/L	2020
IgG3	0,131 g/L	0,2-1,1 g/L	2007
IgA	0,4 g/L	0,83-4,07 g/L	2007
IgM	0,2 g/L	0,34-2,14 g/L	2007

*Autoantibodies*

Lupus anticoagulant	Present		2020
Anticardiolipine IgG	Absent		2020
B2 GP1 IgG	<11 U/mL	<66 U/mL	2020

*Vaccine response*

T-dependent	N/A	Under continuous SCIG	
T-independent	N/A	Under continuous SCIG	

*Peripheral blood mononuclear cell immunophenotyping*

<u>B cells</u>	2%	6-19%	2019
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## Memory B cells

IgD+ CD27+	4.0%	6,7%-25,0%	2014
IgD- CD27+	1.2%	12,5%-14,0%	2014

## CD21 low B cells

CD21 low B cells	3.67%	1.63%-2,56%	2014
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T cells

CD4/CD8 ratio	0.62	1-4,4	2014
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## CD4

Naïve	7.7%	23,9%-46,8%	2014
CM	28,6%	31,8%-50,1%	2014
EM	41,2%	6,6%-18,4%	2014
EMRA	14,8%	0,6%-3,3%	2014

## CD8

Naïve	6.4%	13,8%-33,3%	2014
CM	2,3%	3,9%-11,1%	2014
EM	38,9%	13,0%-27,1%	2014
EMRA	52,4%	33,1%-55-1%	2014

Intracellular cytokine staining after PMA/ionomycin stimulation

## CD4

IFNγ	62,70%	16,93%-22,38%	2020
TNF	77,90%	37,53%-47,63%	2020
IL-2	22,60%	20,55%-30,05%	2020
IL-13	0,22%	0,09%-0,19%	2020
IL-17A	0,97%	1,41%-2,60%	2020
IL-21	10,40%	1,84%-6,52%	2020

## CD8

IFNγ	89,90%	45,25%-72,83%	2020
TNF	57,50%	29,18%-45,68%	2020

NK cells

CD56hi	0,19%	0,49%-0,79%	2014
CD16+ CD56+	19,90%	10,18%-15,40%	2014
CD16+ CD56-	4,43%	0,13%-0,24%	2014

Monocytes

	11,93%	25,63%-29,83%	2014
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cDCs

	0,38%	0,84%-1,19%	2014
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pDCs

	0,21%	0,40%-0,66%	2014
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iNKTs/MAITs

	0,21%	2,45%-3,30%	2014
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